With the Compliments of the Author.

COLOBOMA LENTIS.

BY

ALBERT G. HEYL, M.D.,

OF PHILADELPHIA.

[Reprinted from the Report of the Fifth International Ophthalmological Congress held in New York, September, 1876.]

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COLOBOMA LENTIS.

I PROPOSE in the following paper to offer some remarks upon the congenital irregularity of structure known as coloboma of the crystalline lens. Cases of the kind are clinical curiosities, and so far as my knowledge goes no extended account of them exists. Independent of their mere variety, however, they bear directly upon the process of development of the lens in the embryo, and, mainly with this idea in view, I have been led to give a description of three cases of the kind recently under my own observation, and at the same time to gather and weave in the meagre details relating to the subject scattered far and wide over the field of ophthalmic literature. The history of Case I. is as follows:—

CASE I.—Friedrich Hess, aged fifty-one, applied for relief for impaired vision at the Eye and Ear Department of the Philadelphia Dispensary. The sight of the left eye had been lost for some time; that of the right has been rapidly failing. Before this loss of sight, he had found no difficulty in attending to his occupation, that of a peddler. Examination of each eye gave the following result, the appendages of both eyes being normal:—

R. E., V. Sn. $\frac{5}{200}$; with $-11 = \frac{20}{200}$. Iris of medium gray color, pupil of normal appearance, excepting below, where it opens into a large cleft in the iris tissue; this coloboma iridis does not extend quite to the ciliary margin, but is separated from it by a small fragment of iris tissue. This span of tissue divides the coloboma into two portions: a larger communicating directly with the pupil, and a smaller about the size of a pin-head, completely surrounded by iris tissue. No tremulous motion of the iris perceptible. After instillation of atropine a slow and but partial dilatation took place. The upper part of the iris scarcely responded to the atropine, while the effect upon the lower portions was quite noticeable.

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Examination with the ophthalmoscope now revealed very clearly the peculiar condition of the lens, to which your attention is particularly directed. Directly behind the pupillary space a portion of the edge of the lens was seen, not presenting the well-known rounded contour, being perfectly straight and horizontal; on the nasal side and behind the iris, the edge terminated in a small rounded protuberance, which in turn on its nasal aspect touched the equator. Further examination revealed an enormous coloboma of the choroid, extending beyond the optic nerve; the edge of the disk not discernible; the sclerotic in the position of the coloboma bulged outwardly to a considerable extent. One retinal artery accompanied by two veins seen in the upper part of the fundus, but no retinal vessels could be found below; below the optic disk several vessels pierced the sclerotic, which, judging from their size, must have been the short posterior ciliary arteries; one of the long posterior ciliary arteries was traced far forward, and one of the vortex veins carrying the return-blood was seen piercing the sclerotic at the equator of the ball. Whether a cleft existed in the ciliary body, could not be ascertained; refraction through the lens was myopic to the extent of one-seventh. Hypermetropia of one-eighth existed where the lens was deficient. These measurements are only approximative, owing to the cloudiness of the media and other sufficient reasons.

L. E. Pupil occluded; lens completely opaque, having undergone calcareous degeneration. Coloboma of iris, lens, and choroid existed, but, owing to the occlusion of the pupil, it was impossible to make a further examination.

Case II. may be briefly described as follows: George Dowd, aged twenty, applied for treatment of a chronic catarrh of the middle ear. A coloboma of the iris was noticed in the left eye, and led to a more careful examination, with the hope that possibly a defect might be found in the lens. The result of the examination was as follows: L. E., V. = $\frac{20}{100}$, not improved by glasses. A complete coloboma of the iris, *i. e.*, one extending to the ciliary margin, existed. Corresponding to this, there was a slight deficiency in the periphery of the lens, and presenting, in place of a straight outline, one made up of several sinuosities, taking, on the whole, a horizontal direction; the deficiency was so small that it was with difficulty made out; a large coloboma of choroid existed, not including, however, the optic disk; some opaque nerve-fibres were noted extending downward from the optic disk. The refraction was strongly hypermetropic, no cloudiness of the media existed, no dislocation of the lens. R. E., V. = $\frac{20}{20}$. A slight grade of hypermetropia existed; no abnormal appearances were noted, although no dilatation of the pupil was made, and therefore it is impossible to speak with absolute certainty about the condition of the lens.

Case III.—Mary Adams, aged fifty-two, presented herself at the dispensary for some relief for her eyesight. Her condition was as follows: R. E., no light perception; myopia of one-fifth present. L. E., V. = $\frac{20}{30}$. Emmetropia.

A large coloboma of the iris, of unusual form and position, existed; it was elliptical in shape and situated entirely in the inner half of the iris, the latter being conceived of as divided into two portions by a vertical plane; almost the whole inner half of the iris was absent, a narrow edging of iris tissue skirting the ciliary margin. The whole pupil was involved in the coloboma, and, had it been perfect in shape, would have appeared displaced inward. There was no dislocation of the lens and no evidence of any opacities. By causing the patient to look downward, a small deficiency was discovered at the periphery and extending into the substance of the lens to the extent of perhaps a millimetre; its direction in the main was horizontal; its outline not perfectly straight, but uneven at one or two points, resembling closely the last case. The coloboma was situated in the lower half of the lens; no evidence of coloboma was seen in the interior of the eye. The blindness in this eye was accounted for by the existence of glaucoma absolutum. The L. E. did not present on examination, as far as could be determined, any evidence of coloboma.

In order to gain a clear and comprehensive insight into these cases, I propose to speak a little in detail of the four salient clinical features which they present.

1. *The Outline of the Lens.*—The main peculiarity about this in my first case was the straight horizontal edge already described.

R. Wagner[1] describes a similar case in which the lens appeared as if a segment had been cut off from it, leaving a clearly cut straight edge; the lens outline in my second case

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[1] Ammon's Zeitschrift, Bd. iii., S. 288.

was somewhat analogous, in that, although presenting several sinuosities, it took on the whole a horizontal direction. Bowman has reported a similar case.[1] This, however, is not the usual condition; as a rule, the edge of the lens, instead of being straight, is curved, the convexity being upward. Thus, Bowman has reported another case[2] in which "the border approached a triangular outline in a way to give an idea of the trilinear arrangement of the segments." Jaeger's case,[3] the same from which the beautiful plates of coloboma of the choroid were taken, was similar, the lens being indented to the extent of about two millimetres, giving one the idea of the under part of a saddle (sattelförmig eingekerbt). Arlt described a case[4] in which a slight notch was discovered in the edge of the lens opposite a raphe in the corpus ciliare. the latter being due to imperfect development. Another case[5] is reported by the same author, in which the deficiency, while being quite shallow, was more extended than in the last instance, the lens being "flach eingekerbt." Schiess has described a case,[6] almost exactly similar, the only apparent difference being in the amount of deficiency, which was still greater than in the last-mentioned case, involving about one-fourth of the equator of the lens. Bresgen has lately described a case[7] in which coloboma of the lens was found on each eye, and, judging from the report of it, resembling very closely in outline Schiess' case.

2. *The amount of deficiency* next claims our attention; this varies from a slight indentation to about one-quarter of the lens substance; in no recorded case has it attained to more than the latter amount. Total absence of the lens has been apparently observed in microphthalmic eyeballs, as in the case described by Seiler and Von Ammon;[8] but cases of this

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[1] Ophthalmic Hospital Reports, vol. v., I., p. 72.

[2] Ibid., p. 2.

[3] Ophthalmoskopischen Hand-Atlas, S. 147.

[4] Krankheiten des Auges, Bd. ii., S. 128.

[5] Ibid., S. 129.

[6] Klinische Monatsblätter, 1871, S. 99.

[7] Knapp's Archiv., Bd. iv., II., S. 227.

[8] *Vide* Ruete, Augenheilkunde, Braunschweig, 1855, S. 635.

kind belong to a different category from the one under discussion, and it is very probable, also, that careful examination of these cases would have revealed either the lens or its analogue occupying an unusual position. Aphakia congenita, resulting from defective development, has never been observed in eyeballs at all well developed, and probably never will, when the part played by the lens, as Becker[1] has remarked, in the development of the eye, is remembered. Be this as it may, it is remarkable to find that, in cases of coloboma of the lens, already given, the deficiency never involves the centre of the lens, its anterior or posterior pole, nor the lens substance in their immediate vicinity; on the other hand, that always the equator with adjoining lens substance is involved, and further, that the coloboma extends completely through the lens from before backward. It may be remarked, in this connection, that sometimes in these cases not only does the lens appear deficient at a particular point, but also to be imperfectly developed in all its meridians, so that, even when the eyeball is fully formed, the lens itself is small; thus, in Bowman's cases No. I.[2] and No. V.[3] Again let us notice

3. *The position of the deficiency*, viz., in the inferior half of the lens, the latter being conceived of as divided into two portions by a horizontal plane passing through its centre. This was true of all the cases already cited, excepting Schiess',[4] in which a small portion of the defect was in the superior half, by far the greater part, however, below the dividing line. The common direction of the coloboma was downward, the lens being supposed to be in normal position; of course, a luxation of the lens would alter the direction of the coloboma. In Schiess' case we must notice again an exception; here, without any evidence of luxation, the defect pointed downward and outward. Once more let us notice

4. *Coloboma* of other structures of the eye is not unfrequently an accompaniment of coloboma of the lens; thus, in Wagner's

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[1] Handbuch der Augenheilkunde, Graefe and Saemisch, Bd. v., I., S. 229.

[2] Ophthalmic Hospital Reports, vol. v., I., p. 72.

[3] Ibid., p. 2.

[4] Klinische Monatsblätter, 1871, S. 99.

case,[1] coloboma of the iris, but not of the choroid or retina, existed. In my own cases the choroids, irides, and doubtless the retinæ, were affected.

In Bowman's cases [2,3] no reference is made to it. Arlt's case No. I.[4] was characterized by coloboma of the iris, choroid, and probably retina; a "raphe" existed likewise in the corpus ciliare. Arlt's second case[4] was accompanied by coloboma of the iris, corpus ciliare, and possibly of the choroid. In Schiess' case[5] no mention is made of this condition; the iris, as is evident from the sketch of the case, was perfect; whether any trace of coloboma was to be found in the fundus does not appear. In Jaeger's case,[6] coloboma of the iris and choroid coexisted. In Bresgen's case,[7] coloboma of the other structures did not exist. The same may be said of a case observed by Becker,[8] and also of one communicated by Hirschberg.[9]

From the preceding, it may then be seen that coloboma of the lens is a condition which presents at the place of defect an edge, not rounded as in the normal condition, but either straight in the horizontal direction, or incurved; that the amount of deficiency varies from a slight indentation to about one-quarter of the lens substance, the centre of the lens, its poles, and the lens substance, in the immediate vicinity, being uninvolved; that the lens sometimes, in addition to the defect, is imperfectly developed in all its meridians; that the deficiency is always in the interior half, and almost, without exception, entirely so; and, finally, that coloboma of the other ocular structures frequently coexists, but very frequently no trace of it can be found.

Bearing, then, these clinical features in mind, let us endeavor to trace, as far as possible, the connection between them and some points in the lens development, for, evidently, the coloboma lentis is due to an arrest of this formative process.

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[1] Ammon's Zeitschrift, Bd. iii., S. 288.

[2] Ophthalmic Hospital Reports, vol. v., I., p. 72.

[3] Ibid., p. 2.

[4] Krankheiten des Auges, Bd. ii., S. 128.

[5] Klinische Monatsblätter, 1871, S. 99.

[6] Ophthalmoskopischen Hand-Atlas, S. 147.

[7] Knapp's Archiv., Bd. iv., II., S. 227.

[8] Handbuch der Augenheilkunde, Graefe and Saemisch. Bd. v., I., S. 230.

[9] Ibid.

It will suffice for my purpose merely to mention some of the stages of this process without entering upon the complicated details; those who may wish to examine the subject more closely are referred to the articles by Manz,[1] and also by Arnold,[2] in the "Handbuch der Augenheilkunde," edited by Graefe and Saemisch, or Arnold's monographs.[3] Notice then:

1. The lens is derived from the external layer of the blastodermic membrane, and at a very early period of development separates from it; it then appears as a rounded body, surrounded by a lighter-colored tissue, which is a portion of the middle layer of the blastodermic membrane. This rounded body presents upon section a central lighter-colored portion, composed of rounded bodies and a darker peripheral portion composed of a granular material.

2. The contents of this body undergo various changes, by virtue of which the peripheral portion is found to be composed of elongated nucleated cells, the long diameter being perpendicular to the external surface of the lens, and the inner extremity pointing toward the centre. It is to these peripheral cells that I call your special attention, because it is from them that the lens structure is mainly formed. They do not, however, all contribute in the same degree to the formation of the lens; the cells anterior to the equator are transformed into the epithelium of the anterior capsule; the cells posterior to the equator become transformed into the lens fibres. This latter transformation does not take place in these cells at the same time; the cells grouped about the posterior pole are the first to show signs of increase in length, while those situated more toward the equator seem to remain stationary; so that, if a section be made in the meridian of the lens at this period of development, we shall see a cellular mass projecting from

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[1] Handbuch der Augenheilkunde, Graefe and Saemisch, Bd. ii., I.

[2] Ibid., Bd. i., S. 288.

[3] Entwickelungs Geschichte des Auges, Julius Arnold.

the region of the posterior pole into the central portion of the lens, and causing this hitherto rounded space to assume a meniscoid shape. Afterward, the cells at the side begin to lengthen, and are finally converted into lenticular fibres. It is this latter phase of development which Becker[1] has so well observed.

3. The fœtal blood-supply of the lens needs also a brief notice. The lens during its intra-uterine development is surrounded by a close network of vessels which originate as follows: Those covering the anterior surface are off-shoots from the vessels of the so-called "cerebral plates," while those on the posterior surface are derived from the hyaloid artery; this vessel may be seen at an early period near the posterior pole of the lens, and from this point off-shoots are developed, which gradually cover the posterior surface. This last observation of Arnold explains sufficiently why the cells grouped about the posterior pole should be the first to elongate, as evidently the first signs of growth will be noticed at the place where the bloodvessels first appear; and, as the latter extend on every side toward the equator of the lens, so may we expect to see the remaining cells undergoing the same process.

These statements being clearly understood, we are now in a position to compare them with the clinical features already given, and ascertain something at least about the abnormal processes of development which give rise to this condition of coloboma of the lens. At the outset it must be evident that we cannot conclude that coloboma of the lens arises from any cause acting during the earliest stage of lens development; the constancy of the position of the defect, the position itself, the symmetry of the existing portions of the lens, forbid this supposition. Nor can we look upon this defect as the result of imperfect closure of tissue in a median line, as in the cases of retinal coloboma; the mode of development already described renders some other explanation necessary. Nor, finally, can we conclude that the trilinear arrangement of segments has anything to do with the causation of this defect, for the

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[1] Graefe's Archiv für Ophthalmologie, Bd. ix., II., S. 1–42.

defect is not limited to either of the segments, nor does the mode of development warrant such a supposition.

The second statement, viz., that signs of growth are first noticed in the cells grouped about the posterior pole, and afterward in those nearer the equator, gives us a clue to the solution of the problem. It must be evident that, if the development proceed normally in the first-named position, and cease in the second, we shall have a lens perfect as to its centre, defective as to its peripheral portions. Let me here say that it is not asserted that the development of these two portions is entirely independent, the one of the other; very far from it. While, theoretically, we may have a lens in which only a central portion including the poles exists, we cannot possibly have one in which this is absent and the more peripheral portion present. This finds an explanation in the third statement, viz., regarding the bloodvessel arrangement about the posterior surface of the lens. Should the principal vessel already alluded to fail, there will, of course, be no offshoots, and we shall find only rudimental remains of a lens; should some of its offshoots fail, we shall expect to find a corresponding failure in development of the peripheral portion. The extent of the defect and the character of its outline will naturally vary in different cases.

This solution, then, is based upon the vascular supply about the posterior surface of the lens, and requires that, in cases of simple coloboma—

1. The defect must extend completely through the lens from before backward.
2. The central fibres must be intact.
3. The extent of defect and character of outline must vary.

I need hardly say that the clinical features of coloboma lentis, already given, coincide entirely with these theoretical statements.

Let us now turn our attention to one more point. Theoretically, coloboma of the lens may occur with equal facility at any point of the equator, or even, as already hinted, involve the whole equatorial region; clinically, as may be remembered, the lower half of the lens is always involved, the upper half never, unless in connection with the lower, and then very slightly. We notice at once the frequent presence of colobo. mata of other ocular structures occupying the same relative position, but, for patent reasons, cannot expect to find any direct causal connection between them; much rather must we suppose that both of these conditions are the effect of one cause. The probability would seem to be, that the delayed closure, or the failure to close on the part of the fœtal cleft, must exercise the leading influence in the production of this condition.

The manner in which this process affects the development of the bloodvessels surrounding the lens is not clear to us, and cannot be made so until exact investigation shall have shed more light upon the whole subject of coloboma oculi. Many suggestions have doubtless occurred to all who have given the subject careful thought; but it has seemed to me that to follow them out would only result in drifting into a vortex of useless hypotheses. In my opinion, then, we must at present be content with seeing how this anomalous condition may be produced, providing its exciting cause come into play, the latter being not definitely known to us. * * * * *

In endeavoring, then, to weld these scattered links of observations into the great chain of ophthalmic science, I have taken the eighteen instances of the defect, all that I could find after careful and extended search, and picked out the salient characteristics. I have then detailed as briefly as possible some points of the lens development, and endeavored to show how an arrest of development at a particular period would cause such a condition of affairs as coincided completely with these salient clinical features. Of course, a paper based upon eighteen cases forms but a contribution to the subject; we need, to complete our knowledge of this defect, a larger number of carefully reported cases, accurate investigation both microscopic and macroscopic, and a more definite knowledge regarding the ultimate causes producing coloboma oculi. A few additional remarks have also been added regarding the associated pathological changes, because they have been brought out in the study of the condition, and may serve as a nucleus for further investigations on these points.